FACTSHEET

Veterinary Services

United States Department of Agriculture

Animal and Plant Health Inspection Service

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Bovine Spongiform Encephalopathy

Bovine spongiform encephalopathy (BSE), widely referred to as “mad cow disease,” is a chronic degenerative disease affecting the central nervous system of cattle. The disease was first diagnosed in 1986 in Great Britain.

BSE has had a substantial impact on the livestock industry in the United Kingdom. The disease also has been confirmed in domestic cattle in Belgium, France, Ireland, Liechtenstein, Luxembourg, the Netherlands, Portugal, and Switzerland. The U.S. Department of Agriculture’s (USDA) Animal and Plant Health Inspection Service (APHIS) is enforcing import restrictions and is conducting surveillance for BSE to ensure that this serious disease does not become established in the United States.

Clinical Signs

Cattle affected by BSE experience progressive degeneration of the nervous system. Affected animals may display changes in temperament, such as nervousness or aggression, abnormal posture, incoordination and difficulty in rising, decreased milk production, or loss of body weight despite continued appetite. Affected cattle die. The causative agent of the disease is not completely characterized, and there is neither any treatment nor a vaccine to prevent the disease.

The incubation period (the time from when an animal becomes infected until it first shows disease signs) is from 2 to 8 years. Following the onset of clinical signs, the animal’s condition deteriorates until it either dies or is destroyed. This process usually takes from 2 weeks to 6 months. Most cases in Great Britain have occurred in dairy cows between 3 and 6 years of age.

Currently, there is no test to detect the disease in a live animal; veterinary pathologists confirm BSE by postmortem microscopic examination of brain tissue or by the detection of the abnormal form of the prion protein. BSE is so named because of the spongy appearance of the brain tissue of infected cattle when sections are examined under a microscope.

History

From November 1986 (when BSE was first identified as a separate disease entity) until July 2, 1999, 175,065 head of cattle in more than 34,800 herds were diagnosed with BSE in Great Britain. The epidemic peaked in January 1993 at almost 1,000 new cases reported per week. Agricultural officials in Great Britain have taken a series of actions to eradicate BSE, including making BSE a notifiable disease, prohibiting the inclusion of mammalian meat-and-bone meal in feed for all food-producing animals, prohibiting the inclusion of animals more than 30 months of age in the animal and human food chains, and destroying all animals showing signs of BSE and other animals at high risk of developing the disease.

As a result of these actions, the rate of newly reported cases of BSE is decreasing. Currently, approximately 60 new cases are found in Great Britain per week.

Epidemiology

There are different scientific hypotheses concerning the origins of BSE. Epidemiologic data suggest that BSE in Great Britain may have been caused by feeding cattle rendered protein produced from the carcasses of scrapie-infected sheep. Scrapie has a long incubation period—up to 60 months—and has been endemic in Great Britain for centuries. An alternate theory is that BSE had existed in undetectable levels in the British cattle population prior to 1986. The practice of using products such as meat-and-bone meal as a source of protein in cattle rations has been common for several decades. Changes in rendering operations in the early 1980’s may have played a part in the appearance of the disease and the large number of cases that developed.

There is no evidence that BSE spreads horizontally, i.e., by contact between unrelated adult cattle or from cattle to other species. Limited research suggests that maternal or vertical transmission may occur at a very low level. This low

level most likely would not perpetuate the epidemic under British farming conditions. Research continues in this area.

BSE is classified as a transmissible spongiform encephalopathy (TSE). The agent responsible for BSE and other TSE's is smaller than the smallest known virus and has not been completely characterized. There are three main theories on the nature of the agent: (1) the agent is a virus with unusual characteristics, (2) the agent is a prion—an exclusively host-coded protein that is modified to a partially protease-resistant form after infection, and (3) the agent is a virino—a small, noncoding regulatory nucleic acid coated with a host-derived protective protein. The BSE agent is extremely resistant to heat and to normal sterilization processes. It also does not evoke any detectable immune response or inflammatory reaction in host animals.

In cattle naturally infected with BSE, the BSE agent has been found only in brain tissue, in the spinal cord, and in the retina. Additional studies have identified BSE infectivity in the distal ileum, bone marrow, dorsal root ganglion, and trigeminal ganglion of calves that had been fed brain material from BSE-infected animals.

The presence of the BSE agent in tissues is determined by inoculating animals, usually mice, with material believed to be infected with BSE. Mouse inoculation studies take a long time (up to 700 days) to detect the agent, and failure to identify it in tissues may indicate either true absence of the agent or simply the limited sensitivity of current diagnostic methods.

Related Diseases

The TSE family of diseases includes scrapie, which affects sheep and goats; transmissible mink encephalopathy; feline spongiform encephalopathy; chronic wasting disease of deer and elk; and kuru, both classical and variant Creutzfeldt–Jakob disease, Gerstmann–Straussler–Scheinker syndrome, and fatal familial insomnia, five rare diseases in humans. TSE's have also been reported in Europe in captive wild ruminants, cats, and monkeys. The occurrence of TSE's in captive wild animals is believed to have resulted from BSE-contaminated feed.

On March 20, 1996, the United Kingdom's Spongiform Encephalopathy Advisory Committee (SEAC) announced the identification of 10 cases of vCJD. These 10 cases differed from other routinely diagnosed cases of classical CJD in that they shared an early age at onset of symptoms, an unusual clinical course with psychiatric problems, an overall prolonged duration of illness, and, under microscopic examination, different lesions in brain tissues.

SEAC concluded that, although there was no direct scientific evidence of a link between BSE and vCJD, based on current data and in the absence of any credible alternative, the most likely explanation at that time was that the cases were linked to exposure to BSE before the introduction of a specified bovine offal (SBO) ban at slaughter in 1989. The SBO ban excluded from human consumption brain, spinal cord, and other organs with potential BSE infectivity. As of July 1999, 45 probable cases of vCJD had been identified.

It is important to clarify the difference between classical CJD and vCJD. Classical CJD occurs each year at a rate of 1 to 2 cases per 1 million people throughout the world, including in the United States and other countries where BSE has never occurred and among vegetarians and meat eaters alike. Classical CJD occurs sporadically and is not linked to BSE. According to the U.S. Department of Health and Human Services' Centers for Disease Control and Prevention (CDC), no cases of vCJD have been found in the United States.

Research published in October 1997 found evidence to further link vCJD to BSE. Investigators in the United Kingdom found that BSE and vCJD are of the same "strain." In addition, classical CJD and known scrapie strains were not similar to vCJD or BSE.

USDA Actions in Response to BSE

The United States has one of the most aggressive BSE surveillance programs in the world. BSE has not been diagnosed in the United States, and USDA has worked proactively to keep it that way. In cooperation with USDA's Food Safety and Inspection Service (FSIS), APHIS has taken stringent measures in prevention, education, surveillance, and response.

To prevent BSE from entering the country, since 1989 APHIS has prohibited the importation of live ruminants from countries where BSE is known to exist in native cattle. Other products derived from ruminants, such as fetal bovine serum, bonemeal, meat-and-bone meal, bloodmeal, offal, fats, and glands, are also prohibited from entry, except under special conditions or under USDA permit for scientific or research purposes.

On December 12, 1997, APHIS stopped the importation of all live ruminants and most ruminant products, including meat-and-bone meal, offals, glands, etc., from Europe until APHIS scientists can assess the disease risk and evaluate surveillance activities in individual countries. In addition to the countries already known to have BSE, these new restrictions apply to Albania, Austria, Bosnia–Herzegovina, Bulgaria, Croatia, Czech Republic, Denmark, Federal Republic of Yugoslavia, Finland, Germany, Greece, Hungary, Italy, the former Yugoslavian republic of Macedonia, Norway, Poland, Romania, Slovak Republic, Slovenia, Spain, and Sweden.

APHIS educates veterinary practitioners, veterinary laboratory diagnosticians, industry, and producers on the clinical signs and pathology of BSE.

APHIS leads an ongoing, comprehensive, interagency surveillance program for signs of BSE in the United States. APHIS veterinary pathologists and field investigators have received training from their British counterparts in diagnosing BSE. FSIS antemortem inspection procedures include identifying animals with central nervous system conditions. Animals identified with such conditions are prohibited from slaughter and referred to APHIS for review. APHIS' surveillance program is based on laboratories' histopathologically examining brains from high-risk cattle, i.e., adult cattle that exhibit clinical signs of a neurological disorder. In addition, using a technique called immunohistochemistry, scientists test brain tissues for the presence of the protease-resistant prion protein. Samples are submitted from nonambulatory cattle and from neurologically ill cattle and rabies-negative cattle identified on the farm, at slaughter, or at veterinary diagnostic laboratories and teaching hospitals. As of August 20, 1999, more than 8,400 brains from the United States and Puerto Rico have been examined with no evidence of BSE or other TSE detected.

APHIS also monitors the remaining cattle imported from Great Britain, Belgium, and other European countries (before the bans on imports from those countries went into effect). As of August 25, 1999, of the 496 cattle imported from Great Britain and Ireland between 1981 and 1989, 10 animals are still alive. The animals are quarantined and observed regularly. To date, no evidence of BSE has been detected. APHIS continues to attempt to purchase the 10 live animals for diagnostic research purposes. The 2 Belgian cattle imported in 1996 and 34 European cattle imported in 1996–97 that are still alive are currently under quarantine, and APHIS is attempting to buy these animals as well.

APHIS has also drafted an emergency response plan to be used in the event that BSE is introduced into the United States. In addition, APHIS' TSE Working Group monitors and assesses all ongoing events and research findings regarding TSE's. APHIS continually revises and adjusts prevention and diagnostic measures as it receives new information and knowledge.

As an additional preventative measure, APHIS supports the Food and Drug Administration's (FDA) regulation (effective August 4, 1997) prohibiting the use of most mammalian protein (with certain exceptions) in the manufacture of animal feeds given to ruminants. In addition, the final regulation also requires process and control systems to ensure that ruminant feed does not contain the prohibited mammalian tissues.

Getting the Word Out

As part of its increased surveillance activities, APHIS is continuing an education effort to inform U.S. cattle producers and veterinarians about this disease. Numerous briefings have been held for industry groups. In addition to press releases and factsheets, a videotape on BSE and an information packet were distributed to all APHIS field offices, State veterinarians, extension veterinarians, colleges of veterinary medicine, and industry groups.

For additional information, contact

USDA, APHIS, Veterinary Services
Emergency Programs
4700 Riverdale Road, Unit 41
Riverdale, MD 20737–1231
Telephone: (609) 259–5825 or (301) 734–8073

For information about importing animals or animal products, contact

USDA, APHIS, Veterinary Services
National Center for Import/Export
Animals Program
Telephone: (301) 734–8170
Products Program
Telephone: (301) 734–7885

For public health information, contact CDC at (404) 639–3091. For food safety information, contact FSIS at (202) 205–0293 or call the USDA's Meat and Poultry Hotline at (800) 535–4555. For more information about the ruminant feed ban, call FDA's Consumer Hotline at (800) 532–4440.

Current information on animal diseases and suspected outbreaks is also available on the Internet. Point your Web browser to http://www.aphis.usda.gov to reach the APHIS home page.
